

1. The hematologic features of congenital hypoplastic anemia (Diamond-Blackfan Anemia) include all the following **EXCEPT**

- ☒ A. normocytic anemia
- B. reticulocytopenia
- C. deficiency or absence of red blood cell bone marrow precursors
- D. elevated fetal hemoglobin (Hb F)
- E. elevated serum iron levels

2. The **MOST** specific test that is helpful to differentiate Diamond-Blackfan anemia from transient erythroblastopenia of childhood is

- A. hemoglobin electrophoresis
- B. reticulocytes count
- ☒ C. erythrocyte adenosine deaminase (ADA) enzyme level assay
- D. bone marrow examination
- E. mean corpuscular volume (MCV)

3. Anemia and reticulocytopenia that occur in the 2nd half of infancy period is **LEAST** likely due to

- A. congenital hypoplastic anemia (Diamond-Blackfan anemia)
- B. transient erythroblastopenia of childhood
- ☒ C. a protracted, prolonged course of the anemia of hemolytic disease of the newborn
- D. aplastic crises complicating various types of chronic hemolytic anemias
- E. Fanconi anemia

4. The main stay of treatment for congenital hypoplastic anemia is

- ☒ A. corticosteroids
- B. androgen
- C. antithymocyte globulin (ATG)
- D. fully matched-related stem cell transplantation
- E. fully matched-unrelated stem cell transplantation

5. All the following are true regarding Transient Erythroblastopenia of Childhood (TEC) **EXCEPT**

- A. it is more common than congenital hypoplastic (Diamond-Blackfan) anemia
- B. most of the affected children are older than 12 mo at onset



- C.** Parvovirus B19 infections is a common causative agent
- D. virtually all children recover within 1-2 months
- E. corticosteroid therapy is of no value
6. The valuable diagnostic feature that differentiate anemia of chronic disorder from iron deficiency anemia is
- A. low serum iron
- B.** low or normal serum transferrin
- C. normal bone marrow cellularity
- D. normochromic normocytic RBC
- E. leukocytosis
7. All the following are true regarding physiologic anemia of infancy **EXCEPT**
- A. it is exaggerated in premature infants
- B. the hemoglobin concentration range is between 9-11 g/dL
- C. the hemoglobin concentration rarely falls below 10 g/dL in healthy infants
- D. vitamin E deficiency does not play a role
- E.** iron deficiency is a contributing factor
8. Folic acid deficiency may develop in the following children **EXCEPT**
- A. a term infant fed on goat's milk
- B. an infant fed on unfortified powdered milk
- C.** an infant fed on pasteurized cow's milk
- D. a child with chronic use of phenytoin anticonvulsant
- E. a child with trimethoprim prophylaxis for recurrent urinary tract infection
9. A 1-year-old child has folic acid deficiency since the age of 4 months, the best indicator of this deficiency is
- A. significant increment of mean corpuscular volume MCV
- B. significant fall of reticulocytes count
- C. high level of lactate dehydrogenase LDH
- D.** decrease level of RBC folate
- E. increase number of hypersegmented neutrophils
10. The first laboratory marker in progressive iron deficiency anemia is
- A.** depletion of bone marrow hemosiderin
- B. falling of serum ferritin
- C. decrease of serum iron and increase of the iron-binding capacity
- D. decrease hemoglobin synthesis
- E. hypochromic microcytic anemia
11. An extremely high RBC distribution width (RDW) is **MOST** likely consistent with

- A. iron deficiency anemia
- B. megaloblastic anemia
- C. sideroblastic anemia**
- D. thalassemia
- E. sickle cell anemia

12. The laboratory finding that is **MOST** likely consistent with the diagnosis of spherocytosis is

- A. high reticulocytes count of more than 10%
- B. normal mean corpuscular volume MCV
- C. low mean corpuscular hemoglobin MCH
- D. high mean corpuscular hemoglobin concentration MCHC**
- E. indirect hyperbilirubinemia

13. In hereditary spherocytosis, all the following will resolve postsplenectomy **EXCEPT**

- A. osmotic fragility**
- B. anemia
- C. reticulocytosis
- D. hyperbilirubinemia
- E. hypoplastic/aplastic crises

14. Splenectomy is recommended in all the following conditions **EXCEPT**

- A. a 6-year-old child with hereditary spherocytosis and significant hemolysis
- B. a 7-year-old child with hereditary elliptocytosis and a hemoglobin level of 7 g/dL and corrected reticulocytes count of more than 15%
- C. an 8-year-old child with hereditary stomatocytosis with hemolysis**
- D. a 6-year-old child with thalassemia major with splenomegaly and frequent blood transfusion requirement
- E. a 12-year-old child with chronic immune thrombocytopenia not responding to all modalities of medical treatment

15. The best diagnostic test for paroxysmal nocturnal hemoglobinuria (PNH) is

- A. acidified serum hemolysis (HAM) test
- B. sucrose lysis test
- C. complement assay
- D. flow cytometry**
- E. bone marrow study

16. All the following may occur as a renal manifestation in a child with sickle cell anemia **EXCEPT**

- A. polyuria
- B. hyposthenuria

- C. nephrotic syndrome
- D. hematuria
- ☒ E. acute renal injury

17. The least common infection that may occur in a 6-year-old child with sickle cell anemia is

- A. Pneumococcus sepsis
- B. Haemophilus influenzae meningitis
- C. Salmonella osteomyelitis
- ☒ D. E.coli urinary tract infection
- E. Parvovirus B19 aplastic episode

18. Blood transfusion therapy in a child with sickle cell anemia is indicated in all the following conditions **EXCEPT**

- A. acute chest syndrome
- B. stroke
- C. chronic intolerable pain syndrome
- ☒ D. avascular necrosis of the hip joint
- E. splenic sequestration

19. The best assessment of iron overload for patients with thalassemia major is achieved by

- ☒ A. liver MRI
- B. bone marrow biopsy
- C. serum iron
- D. serum ferritin
- E. total iron binding capacity

20. The criteria for successful bone marrow transplantation in thalassemic patients include all the following **EXCEPT**

- A. age younger than 15-year-old
- B. no hepatomegaly
- ☒ C. no bone deformities
- D. no iron overload
- E. full HLA-matched sibling

21. The best treatment for spinal cord compression at the vertebral canal with neurological symptoms caused by extramedullary hematopoiesis in thalassemic patients is

- A. blood transfusion
- B. splenectomy
- C. iron chelation therapy

- ☒ D. local radiotherapy
- E. vitamin D supplementation

22. You are evaluating a 4-year-old child with β -thalassemia major; he is on chronic transfusion therapy since the age of 1 year; you suspect transfusion-induced hemosiderosis.

Of the following, the organ that is **LEAST** likely to be affected by iron deposition at this time is

- A. liver
- B. pancreas
- ☒ C. heart
- D. pituitary gland
- E. thyroid gland

23. A healthy 5-mo-old boy appears pale. Examination is unremarkable. Lab findings include: Hb 8.1 g/dL; WBC 4,800/mm³; platelets 144,000/mm³; MCV, 111 fL; blood film showed hypersegmented neutrophils; serum B12, 65 pg/mL (low). The infant is vigorously breast fed.

Of the following, the **NEXT** step of the management is

- A. performing Schilling test
- ☒ B. check serum B12 of the mother
- C. parenteral administration of vitamin B12
- D. administration of both folic acid and vitamin B12
- E. transfuse packed RBCs

24. A 9-month-old child with a hemoglobin concentration of 10 gm/dL and marked microcytosis; serum iron and total iron binding capacity are within normal limits; serum ferritin and hemoglobin electrophoresis are also normal.

Of the following, the **MOST** likely diagnosis is

- A. iron deficiency anemia
- B. sideroblastic anemia
- C. β -thalassemia minor
- ☒ D. α -thalassemia trait
- E. anemia due to chronic disease

25. Hereditary intrinsic factor deficiency (HIFD), formerly called congenital pernicious anemia, differs from the typical adult pernicious anemia by all the following **EXCEPT**

- ☒ A. it is prominent at around 3 mo of age
- B. stomach secretes acid normally
- C. stomach is histologically normal
- D. there are no antibodies to parietal cells
- E. there are no associated endocrine disorders

26. Increased incubated osmotic fragility test that is not corrected by the addition of glucose is **MOST** likely suggestive of

- A. hereditary spherocytosis
- B. hereditary elliptocytosis
- C. hereditary stomatocytosis
- D. glucose-6-phosphate dehydrogenase deficiency
- E. pyruvate kinase deficiency**

27. All the following may cause autoimmune hemolytic anemia **EXCEPT**

- A. systemic lupus erythematosus (SLE)
- B. immunodeficiency
- C. mycoplasma pneumoniae infection
- D. lymphoproliferative disorder
- E. penicillin drug administration**

28. All the following are correct regarding treatment of cold agglutinin disease **EXCEPT**

- A. patient should avoid exposure to cold
- B. patient should be treated for underlying disease
- C. plasmapheresis is a modality of treatment
- D. glucocorticoids treatment is a modality of treatment
- E. patient can be treated by splenectomy when there is no response to other modalities of treatment**

29. Fragmentation hemolysis by mechanical injury may be seen in all the following **EXCEPT**

- A. extensive burns**
- B. Kasabach-Merritt syndrome
- C. after cardiac surgery for prosthetic heart valve replacement
- D. thrombotic thrombocytopenic purpura (TTP)
- E. hemolytic uremic syndrome (HUS)

30. All the following conditions may be associated with iron deficiency **EXCEPT**

- A. hookworm infestation
- B. prolonged intravascular hemolysis
- C. celiac disease
- D. congenital heart disease with right to left shunt
- E. prolonged use of isoniazid (INH)**

31. Parvo virus B19 infection may cause transient pancytopenia in

- A. thalassemia major
- B. sickle cell anemia**
- C. hereditary spherocytosis

- D. hereditary elliptocytosis
- E. pyruvate kinase deficiency

32. Fresh frozen plasma (FFP) transfusion is the best treatment for which of the following conditions

- A. hemophilia a
- B. hemophilia b
- C. factor vii deficiency
- D. immunoglobulin replacement therapy
- E. bleeding due to warfarin therapy**

33. Transfusion of fresh frozen plasma (FFP) is efficacious for the treatment of deficiency of all the following coagulation factors **EXCEPT**

- A. factor V
- B. factor X
- C. factor XI
- D. factor XIII**
- E. protein C

34. Fresh frozen plasma (FFP) transfusion/replacement is indicated in all the following conditions **EXCEPT**

- A. a 9-year-old child with chronic liver disease with prolonged clotting times who needs a liver biopsy
- B. a 6-year-old child with protein S deficiency and significant deep venous thrombosis
- C. an 11-year-old child with thrombotic thrombocytopenic purpura (TTP) post allogenic bone marrow transplantation
- D. a 2-day-old preterm neonate with clotting deficiencies to prevent intraventricular hemorrhage**
- E. a 1-week-old term neonate with hemorrhage secondary to vitamin K deficiency

35. The main reason behind gamma irradiation of the blood products is to prevent

- A. transfusion-related bacterial infections
- B. transfusion-related viral infections
- C. graft-versus-host disease (GVHD)**
- D. allergic reactions
- E. transfusion-related acute lung injury (TRALI)

36. CMV-transmitted infection is least likely occurring in transfusion of which of the following blood products?

- A. whole blood
- B. packed RBCs

- C. granulocytes
- D. platelets
- ☒ E. fresh frozen plasma

37. A prolonged thrombin time with normal reptilase time is consistent with

- A. dysfibrinogenemia
- B. afibrinogenemia
- C. factor XIII deficiency
- D. disseminated intravascular coagulopathy (DIC) with high level of fibrin split products
- ☒ E. heparin overdose

38. In hemophilia A, factor VIII level activity should be increased to 100% in

- A. epistaxis
- ☒ B. iliopsoas bleeding
- C. tooth extraction
- D. hematuria
- E. gum bleeding

39. A 9-year-old male child is about to do elective tonsillectomy; preoperative investigations are all normal except for a significant prolonged partial thromboplastin time PTT; there was no previous bleeding after circumcision and emergency appendectomy.

Of the following, the **MOST** likely diagnosis is

- A. hemophilia A
- B. hemophilia B
- C. hemophilia C
- ☒ D. prekallikrein deficiency
- E. factor XIII deficiency

40. Spontaneous intracranial hemorrhage is more likely to occur in

- ☒ A. factor VII deficiency
- B. factor VIII deficiency
- C. factor IX deficiency
- D. factor XI deficiency
- E. factor XIII deficiency

41. Systemic amyloidosis may be associated with deficiency of

- A. factor VII
- B. factor VIII
- C. factor IX
- ☒ D. factor X

E. factor XI

42. In which of the following conditions you should use a plasma that is frozen within 24 hours after collection?

- ☒ A. factor V deficiency
- B. factor VII deficiency
- C. factor IX deficiency
- D. factor X deficiency
- E. factor XI deficiency

43. All the following are true regarding factor XIII **EXCEPT**

- A. the half-life is 5-7 days
- ☒ B. PT, PTT, and thrombin time (TT) are prolonged when the factor is deficient
- C. clot solubility test with 5m urea is a screening test for factor XIII deficiency
- D. factor assay is a specific test to detect its level
- E. the hemostatic level is only 2-3%

44. In severe liver disease with prolongation of the bleeding time, the best effective treatment before doing liver biopsy is

- A. intravenous vitamin K
- B. intramuscular vitamin K
- C. oral vitamin K
- D. fresh frozen plasma
- ☒ E. intravenous DDAVP

45. Vitamin K-dependent clotting factors include all the following **EXCEPT**

- ☒ A. factor I
- B. factor II
- C. factor VII
- D. factor IX
- E. protein S

46. A 16-year-old female is found to have multiple bruising and hypochromic microcytic anemia; during her first pregnancy she noticed disappearance of the bruises; she has a past history of uncomplicated emergency appendectomy at the age of 10 year.

Of the following, the drug **MOST** likely beneficial for alleviation of her symptoms is

- A. ferrous sulphate
- ☒ B. desmopressin acetate (DDAVP)
- C. recombinant factor VIII
- D. recombinant factor IX
- E. aminocaproic acid

47. Low platelets count may be encountered in all the following **EXCEPT**
- A. Von Willebrand disease (VWD) type 2 B
 - B. VWD type 2 N**
 - C. Platelet type pseudo VWD
 - D. Bernard Soulier syndrome
 - E. Wiskott Aldrich syndrome
48. Which of the following is **FALSE** about factor V Leiden mutation?
- A. it is the most common inherited risk factor for thrombosis
 - B. heterozygous individuals have less risk for thrombosis than homozygotes
 - C. heterozygotes have an increased risk of arterial thrombosis**
 - D. there is an increased frequency of thrombosis while receiving oral contraceptive agents
 - E. there is an increased risk of recurrent abortions
49. Arterial thrombosis is the least likely cause of
- A. stroke
 - B. a cold and pulseless lower extremity
 - C. renal infarction
 - D. myocardial infarction
 - E. pulmonary embolism**
50. Lupus anticoagulants usually affect the following laboratory test
- A. bleeding time (BT)
 - B. partial thromboplastin time (PTT)**
 - C. prothrombin time (PT)
 - D. thrombin time (TT)
 - E. clot solubility test
51. Which of the following is **FALSE** regarding standard unfractionated heparin therapy?
- A. it acts by enhancing the antithrombin III activity
 - B. it can be used safely during pregnancy
 - C. it should not be given for a child with bacterial endocarditis
 - D. it can be given intramuscularly**
 - E. it needs monitoring by PTT test
52. All the following are true regarding low molecular weight heparin (LMWH) in comparison with unfractionated heparin (UFH) **EXCEPT**
- A. it is more stable
 - B. it is ease to titrate
 - C. it is monitored by activated PTT**

- D. it can be used subcutaneously
- E. it can be used as an outpatient

53. A healthy 5-day-old male neonate develops bruising and melena. The pregnancy, and delivery course were unremarkable. The mother received heparin therapy for recurrent abortions secondary to lupus anticoagulants. The infant is vigorously breastfeeding. Examination reveals only multiple bruises. Lab tests reveals: hemoglobin, 8.4 g/dL; WBC, 8600/mm³; platelets count, 200000/mm³; PTT, 74 seconds; PT, 28 seconds; serum fibrinogen is normal.

Of the following, the **MOST** likely cause of this bleeding is

- A. disseminated intravascular coagulopathy (DIC)
- B. hemophilia
- C. vitamin K deficiency**
- D. heparin therapy
- E. lupus anticoagulants

54. A previously healthy 10-month-old boy develops pallor over 2 weeks. Examination is unremarkable except for pallor. Lab tests includes: hemoglobin, 4.9 g/dL; WBC, 6700/mm³; platelets count, 180000/mm³; MCV, 79 fL; reticulocyte count 0.8%; hemoglobin electrophoresis, normal.

Of the following, the **MOST** likely cause for these findings is

- A. pure red cell aplasia
- B. transient erythroblastopenia of childhood**
- C. thalassemia minor
- D. aplastic anemia
- E. parvovirus infection

55. You are evaluating a 5-year-old male child with multiple ecchymoses distributed all over the extremities and trunk; examination reveal a palpable spleen 3 cm below the left costal margin. Lab findings include: hemoglobin, 11.3 g/dL; white blood cell count, 8700/mm³; platelets count, 21000/mm³.

Of the following, the most proper **NEXT** step is

- A. bleeding time
- B. coagulation profile
- C. platelets aggregation test
- D. abdominal ultrasonography
- E. bone marrow study**

56. Both quantitative and qualitative platelets defects are seen in

- A. generalized vasculitis
- B. Bernard-Soulier syndrome**
- C. Glanzmann's Thrombasthenia

- D. type I von Willebrand disease
- E. uremia

57. Which of the following viruses is more likely to be associated with chronic immune thrombocytopenia?

- A. cytomegalovirus
- B. parvovirus
- C. influenza virus
- D. parainfluenza virus
- E. human immune deficiency virus**

58. Which of the following is least likely to be associated with prolonged thrombocytopenia in children?

- A. immune thrombocytopenia**
- B. systemic lupus erythematosus (SLE)
- C. HIV infection
- D. type 2B von Willebrand disease
- E. Wiskott-Aldrich syndrome (WAS)

59. Which of the following drugs is least likely to cause thrombocytopenia in children?

- A. valproic acid
- B. phenytoin
- C. sulfonamides
- D. trimethoprim-sulfamethoxazole
- E. heparin**

60. Microthrombocytes are **MOST** likely to be seen in

- A. immune thrombocytopenia (ITP)
- B. Bernard-Soulier syndrome
- C. Wiskott-Aldrich syndrome (WAS)**
- D. Glanzmann's Thrombasthenia
- E. type 2B von Willebrand disease

61. Thrombocytopenia that improves with age is **MOST** likely a feature of

- A. congenital amegakaryocytic thrombocytopenia
- B. Bernard-Soulier syndrome
- C. Wiskott-Aldrich syndrome
- D. thrombocytopenia absent radius syndrome (TAR)**
- E. aplastic anemia

62. You are going to explain the role of anti-D therapy to the parents of a 4-year-old boy with acute immune thrombocytopenia (ITP) who has several ecchymotic lesions located over extremities and trunk. There is no mucosal bleeding.

An important statement that should be included during the discussion is

- A. anti-D therapy is more effective than IVIG therapy in acute ITP
- B. anti-D therapy is very effective to all children with ITP
- C. when anti-D therapy is given, monitoring of hemoglobin concentration is recommended
- D. anti-D therapy is used in acute ITP only
- E. anti-D therapy can be used as initial treatment for children with acute ITP

63. A 9-month-old boy appears pale. Examination reveals a palpable liver 3 cm below the right costal margin and palpable spleen 4 cm below the left costal margin. Lab findings include: hemoglobin concentration, 6.4 g/dl; mean corpuscular volume (MCV), 67 fL; and WBC, 15,000/mm³.

Of the following, The **MOST** valuable test to confirm the diagnosis is

- A. blood smear
- B. serum iron, total iron binding capacity, and serum ferritin
- C. hemoglobin electrophoresis
- D. bone marrow study
- E. abdominal ultrasonography

64. You are explaining the risk of sepsis after splenectomy to medical students. An important statement that should be included during this discussion is that risk is especially high in children

- A. older than 5 yr at the time of surgery
- B. with hereditary spherocytosis
- C. with chronic immune thrombocytopenia (ITP)
- D. after trauma
- E. with Hodgkin lymphoma

65. You are evaluating a 7-year-old male child with cervical lymphadenopathy; the **MOST** likely finding that raise your suspicion of malignancy is

- A. matted nodes
- B. fluctuant nodes
- C. fixed nodes
- D. presence of systemic symptoms
- E. no regression in the size of the nodes to its normal within 2 weeks

66. Of the following, the **MOST** accurate reflection of active erythropoiesis of the bone marrow is

- A. hemoglobin concentration

- B. packed cell volume (PCV)
- C. reticulocyte count
- D. mean corpuscular volume (MCV)
- E. reticulocyte index

67. Hematologic disease with upper limb orthopedic congenital abnormalities with presence of thumbs is **MOST** likely seen in

- A. Fanconi anemia
- B. thrombocytopenia with Absent Radii (TAR) Syndrome
- C. congenital Amegakaryocytic Thrombocytopenia
- D. Glanzmann Thrombasthenia
- E. Bernard-Soulier Syndrome

68. Which of the following is not a feature of Wiskott–Aldrich Syndrome (WAS)

- A. thrombocytopenia
- B. eczema
- C. recurrent otitis media and pneumonia
- D. propensity to develop autoimmune disorders
- E. giant platelets by blood smear

69. A 5-year-old child has multiple bruises on her extremities and oral mucosal bleeding of 3 days duration; she had a mild respiratory tract infection 2 weeks before; physical examination was normal apart from multiple ecchymoses and petechiae.

Of the following, the **NEXT** diagnostic step is

- A. complete blood count
- B. prothrombin time
- C. bleeding time
- D. partial thromboplastin time
- E. antinuclear antibody titer

70. Of the following, the **LEAST** likely feature of Bernard-Soulier syndrome is

- A. autosomal recessive inheritance
- B. severe thrombocytopenia
- C. giant Platelets
- D. abnormal ristocetin test
- E. platelet transfusion is the only reliable therapy

71. Each of the following may cause thrombocytopenia **EXCEPT**

- A. heparin therapy
- B. aspirin ingestion
- C. disseminated intravascular coagulation (DIC)
- D. systemic lupus erythematosus (SLE)

E. hemolytic-uremic syndrome (HUS)

72. Which of the following children with acute immune thrombocytopenia is **MOST** likely to have a benefit from splenectomy?

- A. an 8-year-old girl with multiple bruises and platelets count of 10,000/ml
- B. a 6-year-old boy with mild epistaxis and platelets count of 20,000/ml
- C. a 4-year-old boy with subdural hematoma and platelets count of 100,000/ml after receiving IVIG
- D. an 11-year-old boy with severe rectal bleeding and platelets count of 20000/ml not responding to medical treatment
- E. a 13-year-old female adolescent with moderate menstrual bleeding and platelets count of 50000/ml

73. Of the following, the **LEAST** likely feature of acute immune thrombocytopenia (ITP) is

- A. a preceding viral infection
- B. isolated thrombocytopenia
- C. mucocutaneous bleeding
- D. isolated splenomegaly
- E. increased number of megakaryocytes in the bone marrow

74. A 3-year-old male child develops hematoma and bruising of his right hand next day after falling on the ground; the mother stated that her child has a poor wound healing and a history of delayed umbilical separation during the neonatal period.

Of the following, the **MOST** valuable test for this case is

- A. bleeding time
- B. partial thromboplastin time
- C. prothrombin time
- D. thrombin time
- E. clot solubility test

75. A 5-year-old child, with hemophilia A of severe type, presents to the emergency unit with a groin pain after a minor trauma to his back; his blood pressure is 60/30 mm Hg; his pulse rate is 180/min; he holds his right hip in a flexion position with internal rotation.

Of the following, the **NEXT** step in the management of this child is

- A. factor VIII replacement therapy
- B. intravenous 1-deamino-8-d-arginine vasopressin (DDAVP)
- C. factor VIII assay
- D. abdominal ultrasonography
- E. abdominal computed tomography (CT) scan

76. In a child with a recent diagnosis of hemophilia A who developed moderate epistaxis, the aim of the treatment is to increase the level of factor VIII to
- A. 5-20%
 - B. 35-50%
 - C. 55-60%
 - D. 65-70%
 - E. 80-100%
77. Allogeneic bone marrow stem cell transplantation is least likely to be required in
- A. Fanconi anemia
 - B. thrombocytopenia with Absent Radii (TAR) Syndrome
 - C. congenital Amegakaryocytic Thrombocytopenia (CAT)
 - D. Wiskott –Aldrich Syndrome (WAS)
 - E. severe combined immune deficiency
78. Inherited thrombocytopenias include all the following **EXCEPT**
- A. congenital Amegakaryocytic Thrombocytopenia
 - B. thrombocytopenia with Absent Radii (TAR) Syndrome
 - C. Glanzmann's Thrombasthenia
 - D. Bernard-Soulier Syndrome
 - E. Wiskott–Aldrich Syndrome (WAS)
79. Which of the following is the best treatment for a 5-year-old girl with acute immune thrombocytopenia (ITP) and platelets count of 30,000/ml and mild petechial rash all over her body?
- A. intravenous immunoglobulin (IVIG)
 - B. anti-D gamma globulin
 - C. corticosteroids
 - D. platelets transfusion
 - E. no treatment
80. The standard care for **MOST** children with severe hemophilia is
- A. prevention by F VIII replacement therapy to prevent spontaneous bleeding and early joint deformities
 - B. aggressive treatment by F VIII replacement therapy when significant bleeding occurs
 - C. avoid trauma
 - D. avoid aspirin and other NSAID
 - E. avoid violent contact sports

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- 1.(A). The RBCs in Diamond-Blackfan anemia are usually macrocytic for age, but there is no hypersegmentation of neutrophils.
- 2.(C). Erythrocyte adenosine deaminase (ADA) activity is increased in most patients with congenital hypoplastic anemia, a finding that helps distinguish this disorder from acquired transient erythroblastopenia of childhood. Also, because elevated ADA activity is not a fetal RBC feature, measurement of this enzyme is helpful in diagnosing Diamond-Blackfan anemia in very young infants. Hemoglobin F may be increased in 20% of cases with transient erythroblastopenia of childhood at the time of diagnosis.
- 3.(C). The anemia of hemolytic disease of the newborn can have a protracted course, but this usually terminates spontaneously at 5-8 wk of age. Aplastic anemic crises, frequently caused by parvovirus B19 infections, may complicate various types of chronic hemolytic disease, but usually after the first several months of life.
- 4.(A). Corticosteroid therapy is beneficial in three fourths of patients; the mechanism of its effect is unknown. Stem cell transplantation from a related histocompatible donor has a role in children who do not respond to corticosteroids and who have demonstrated a several-year need for RBC transfusions; The survival results for matched-related donors have been very encouraging, but the responses have been much inferior with the use of partially mismatched siblings or matched unrelated donors.
- 5.(C). In rare instances, a prolonged case of apparent TEC may be caused by parvovirus-induced RBC aplasia, occurring in children with hemolytic anemia or congenital or acquired immunodeficiencies.
- 6.(B). The serum iron level in anemia of chronic disorder is low, but without the increase in total iron-binding capacity (serum transferrin) that occurs in iron deficiency. This pattern of low serum iron and low to normal iron-binding protein (serum transferrin) is a regular and valuable diagnostic feature.
- 7.(E). The iron previously stored from the degraded fetal RBCs can be used for hemoglobin synthesis. The supply of stored iron is sufficient for hemoglobin synthesis, even in the absence of dietary iron intake, until approximately 20 wk of age. Unless there has been significant blood loss, iron stores should be sufficient to maintain erythropoiesis early on.
- 8.(C). Goat's milk and powdered milk are deficient with folic acid. Human breast milk, pasteurized cow's milk and infant formulas provide adequate amounts of folic acid. Anticonvulsants may decrease folate absorption. Trimethoprim may affect folate metabolism.
- 9.(D).

- 10.(A). In progressive iron deficiency, a sequence of biochemical and hematologic events occurs. First, the tissue iron stores represented by bone marrow hemosiderin disappear.
- 11.(C). RDW is usually high in IDA, megaloblastic anemia, and sideroblastic anemia, but it is extremely high in the last.
- 12.(D). The MCHC is a measure of cellular hydration status. A high value (36-38 g/ dL) is characteristic of spherocytosis and a low value is commonly associated with iron deficiency.
- 13.(A). After splenectomy, osmotic fragility often improves because of diminished splenic conditioning and less RBC membrane loss, but not resolved.
- 14.(C). Hemolytic anemia may be associated with hereditary stomatocytosis, but splenectomy is not a recommended treatment. Persistent symptomatic thrombocytosis may follow splenectomy if the hemolysis is not eliminated or markedly decreased. Patients have developed a life-threatening tendency toward in situ thrombosis after splenectomy in association with the abnormal adherence of the stomatocytic RBC to vascular endothelium in conjunction with the thrombocytosis.
- 15.(D). Flow cytometry is the best diagnostic test for PNH. With the use of anti-CD59 for RBCs and anti-CD55 and anti-CD59 for granulocytes, flow cytometry is more sensitive than the classic RBC lysis tests in detecting the reduced glycolipid-bound membrane proteins.
- 16.(E). Renal function is progressively impaired by diffuse glomerular and tubular fibrosis leading to chronic, rather than acute, renal shutdown.
- 17.(D). There is an increased incidence of urinary tract infection with bacteremia in young women with sickle cell anemia.
- 18.(D). Blood transfusions usually do not reverse ischemic damage. Even while on chronic transfusion therapy children can still have transient cerebral ischemia as well as avascular necrosis of the hips and shoulders.
- 19.(A). Quantitative liver iron by approved MRI technology is the best indicator of total-body iron stores and should be obtained in chronically transfused patients after chronic transfusion therapy has initiated.
- 20.(C).
- 21.(D). Extramedullary hematopoiesis can occur in the vertebral canal, compressing the spinal cord and causing neurologic symptoms; the latter is a medical emergency requiring immediate local radiation therapy to halt erythropoiesis.
- 22.(C). Iron is initially deposited in the liver. Liver hemosiderosis develops after 1 yr of chronic transfusion therapy and is followed by iron deposition in the endocrine system. This leads to a high rate of hypothyroidism, hypogonadotropic gonadism, growth hormone deficiency, hypoparathyroidism, and diabetes mellitus. After 10 yr of transfusion, cardiac dysfunction secondary to hemosiderosis begins.
- 23.(B). Older children and adults have sufficient vitamin B12 stores to last 3-5 yr. However, in young infants born to mothers with low vitamin B12 stores (breast fed

infants of vegans mothers or having pernicious anemia), clinical signs of cobalamin deficiency can become apparent in the first 4-5 mo of life.

24.(D). The α -thalassemia traits present as a microcytic anemia that can be mistaken for iron-deficiency anemia. The hemoglobin electrophoresis is normal. The diagnosis is confirmed by DNA testing.

25.(A). In HIFD, symptoms become prominent at an early age (6-24 mo), consistent with exhaustion of vitamin B12 stores acquired in utero.

26.(E). Autohemolysis is moderately or markedly increased in pyruvate kinase deficiency, but the addition of glucose does not regularly correct the abnormality as it does in hereditary spherocytosis.

27.(E). Penicillin may cause hemolysis via the "hapten" mechanism (immune but not autoimmune) by binding tightly to the RBC membrane. Antibodies to the drug, either newly or previously formed, bind to the drug molecules on RBCs, mediating their destruction in the spleen.

28.(E). Splenectomy is not useful in cold agglutinin disease.

29.(A). Extensive burns may directly damage the RBCs and result in hemolysis that results in the formation of spherocytes. Blood loss and marrow suppression may contribute to anemia and require blood transfusion; others cause RBC destruction by mechanical injury.

30.(E). Secondary iron deficiency may complicate the intravascular hemolysis because of urinary hemoglobin and hemosiderin iron loss. Cardiovascular defects involving right-to-left shunts interfere with proper oxygenation leading to secondary polycythemia which may cause iron deficiency due to increased RBC demands production. INH therapy may cause secondary sideroblastic anemia with high serum iron and increased transferrin saturation.

31.(B). Parvovirus B19 is classically associated with isolated red cell aplasia, but in patients with sickle cell disease or immunodeficiencies it can result in transient pancytopenia.

32.(E). Transfusion of FFP is no longer recommended for treatment of patients with severe hemophilia A or B or for factor VII deficiency because safer factor VIII, IX, and VII concentrates are available; also it is not indicated for correction of hypovolemia or as immunoglobulin replacement therapy, because safer alternatives exist; An important use of FFP, albeit rare in children, is for rapid reversal of warfarin effects in patients who are actively bleeding or who require emergency surgery (i.e., in whom functional deficiencies of factors II, VII, IX, and X cannot be rapidly reversed by vitamin K).

33.(D). Transfusion of FFP is efficacious for the treatment of deficiencies of clotting factors II, V, X, and XI. Factor XIII and fibrinogen deficiencies are treated with cryoprecipitate. FFP also contains several anticoagulant proteins (antithrombin III, protein C, and protein S), whose deficiencies have been associated with thrombosis.

34.(D). In neonates, Clotting times are prolonged, owing to developmental deficiency of clotting proteins, and FFP should be transfused only after reference to normal values expected for the birthweight and age of the infant. The use of prophylactic FFP

transfusions to prevent intraventricular hemorrhage in premature infants is not recommended.

35.(C).

36.(E). CMV infections may be transmitted by leukocyte containing blood component transfusions; FFP does not transmit such infection because it is acellular component.

37.(E). Unlike the thrombin time, the reptilase time is not sensitive to heparin and is prolonged only by reduced or dysfunctional fibrinogen and fibrin split products.

38.(B).

39.(D). Deficiencies of the contact factors prolong the PTT but are not causes of clinical bleeding. These factors include factor XII, prekallikrein, and high molecular weight kininogen.

40.(A).

41.(D). Systemic amyloidosis may be associated with factor X deficiency owing to the adsorption of factor X on the amyloid protein. In the setting of amyloidosis, transfusion therapy is often not successful because of the rapid clearance of factor X.

42.(A). Factor V is lost rapidly from FFP. It is therefore important to use FFP that is less than 2 mo old. Because the plasma half-life of factor VII is 2-4 hr, therapy with FFP is difficult and often complicated by fluid overload.

43.(B). The usual screening tests for hemostasis are normal in patients with factor XIII deficiency, antiplasmin deficiency, and plasminogen activator inhibitor deficiency.

44.(E). DDAVP (0.3 µg/kg IV) has been found to be effective in shortening the bleeding time and has been used effectively to augment hemostasis prior to liver biopsy.

45.(A). Vitamin K-dependent clotting factors include factors II, VII, IX, X, protein C, and protein S.

46.(B). Von Willebrand disease (VWD) is the most likely cause for this case; menstruating female may present with iron deficiency due to menorrhagia. Because VWF is an acute-phase protein, stress will increase its level; thus, patients may not bleed during appendectomy and childbirth. Bruising symptoms may diminish during pregnancy, because the VWF levels may double or triple during pregnancy.

47.(B). Type 2N VWD is caused by the reduction of factor VIII binding by VWF. This disorder has also been termed autosomal hemophilia. With this variant, platelet interaction with VWF is normal but the 2N VWF binds weakly (or not at all) to factor VIII, resulting in rapid clearance of factor VIII that is weakly complexed to VWF.

48.(C). Individuals who are heterozygous have a 5-7-fold increase in risk of venous thrombosis

49.(E). Pulmonary embolism is a feature of venous thrombosis.

50.(B). The lupus anticoagulant causes a prolongation of the PTT that fails to correct on 1:1 mixing with normal plasma. The antibody is directed against the phospholipid used as a reagent in the PTT.

51.(D).

52.(C). Unlike UFH, which is monitored using the aPTT, LMWH is monitored via the anti-Xa activity.

- 53.(C). Heparin does not cross the placenta, and so it does not affect the baby. DIC causes prolonged PTT, PT, thrombocytopenia and low serum fibrinogen. Hemophilia causes prolonged PTT only. Lupus anticoagulants may cause thrombosis rather than bleeding and usually associated with prolonged PTT because of the presence of antiphospholipids that affect the phospholipid reagent.
- 54.(B). Normal hemoglobin electrophoresis usually excludes thalassemia and Diamond-Blackfan syndrome. Anemia due to parvovirus infection usually occurs in children with hemolytic anemia or immunodeficiency. In aplastic anemia, the anemia is usually macrocytic rather than normocytic.
- 55.(E). Although spleen may be palpable in 10% of children with ITP, bone marrow examination is mandatory when there is an unusual presentation.
- 56.(B). In Bernard-Soulier syndrome there is moderate thrombocytopenia and abnormal ristocetin test.
- 57.(E).
- 58.(A). In 70-80% of children who present with acute ITP, spontaneous resolution of their ITP will occur within 6 mo.
- 59.(E). Common drugs used in pediatrics that cause thrombocytopenia include valproic acid, phenytoin, sulfonamides, and trimethoprim-sulfamethoxazole. Heparin-induced thrombocytopenia is seldom seen in pediatrics.
- 60.(C). WAS is characterized by thrombocytopenia with tiny platelets, eczema, and recurrent infections due to immune deficiency.
- 61.(D). Thrombocytopenia of TAR syndrome frequently remits over the first few years of life.
- 62.(C). The role of IV anti-D in initial therapy of acute ITP is under investigation; the response appears to be somewhat slower than after IVIG; it is effective for Rh+ children; it can be used in acute or chronic ITP.
- 63.(C). β -thalassemia major is the most likely diagnosis; hemoglobin electrophoresis is quite helpful to confirm the diagnosis. Blood smear usually shows hypochromic microcytic RBCs but it is difficult to differentiate between iron deficiency anemia and thalassemia.
- 64.(E). This risk of sepsis is especially high in children younger than 5 yr at the time of surgery. The risk of sepsis is less after splenectomies performed for trauma, RBC membrane defects, and immune thrombocytopenia than when there is pre-existing immune deficiency (Wiskott-Aldrich syndrome, Hodgkin disease) or reticuloendothelial blockade (storage diseases, severe hemolytic anemias).
- 65.(C). A firm fixed node to the surrounding tissue should always raise the question of malignancy, regardless of the presence or absence of systemic symptoms or other abnormal physical findings. Fluctuance suggests abscess formation. Tuberculous nodes may be matted.
- 66.(E). The Reticulocyte production index (RPI, also called a corrected reticulocyte count) is a calculated value used in the diagnosis of anemia. This calculation is

necessary because the raw reticulocyte count is misleading in anemic patients. Reticulocyte index = reticulocyte count × (observed hematocrit/normal hematocrit).

67.(B). TAR is characterized by absent radii with presence of thumbs; this distinguishes from Fanconi anemia in which thumbs are absent.

68.(E).

69.(A).

70.(B). Bernard-Soulier Syndrome is characterized by moderate, rather than severe, thrombocytopenia.

71.(B). Aspirin may affect platelets function rather than the count.

72.(D). Severe life-threatening bleeding not responding to all modalities of medical treatment is an indication for emergent splenectomy.

73.(D). Splenomegaly is present in only 10% of cases of acute ITP.

74.(E). The most likely diagnosis is factor XIII deficiency.

75.(A). Iliopsoas bleeding is life threatening that needs immediate factor replacement therapy.

76.(B). When mild to moderate bleeding occurs, level of factor VIII or IX must be raised to a hemostatic level, in the range of 35-50%.

77.(B). In TAR, thrombocytopenia frequently remits but some cases may require allogenic bone marrow transplantation.

78.(C). Glanzmann's thrombasthenia is characterized by an abnormal platelets function and normal platelets count.

79.(E). Spontaneous recovery occurs in the majority of children with ITP; treatment is required when the platelets count is less than 20,000/ml.

80.(A).